

# Pregnancy outcome and Ebstein's anomaly

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## Abstract

**Background**—Ebstein's anomaly is an uncommon congenital cardiac abnormality that may be associated with cyanosis and arrhythmias. For those female patients with the anomaly who survive to adult life there is little information available about pregnancy, maternal complications, and fetal outcome. This study was designed to address this issue so that these patients can receive appropriate advice and management.

**Methods and results**—Forty two pregnancies in 12 women with Ebstein's anomaly were studied. The mothers' cardiac lesions were assessed on the basis of symptoms, the presence of cyanosis or arrhythmia, and by echocardiographic grading of severity. In the absence of important maternal cyanosis or arrhythmia, pregnancy was well tolerated. Neonatal outcome was good though there was an increased risk of prematurity and dysmaturity in the babies born to mothers with cyanosis.

**Conclusions**—This study indicates that women with Ebstein's anomaly who reach child-bearing age can be advised that pregnancy is likely to be well tolerated with good fetal outcome. Maternal arrhythmia or cyanosis are indications for closer maternal and fetal observation.

In 1866, Wilhelm Ebstein, only five years after his graduation from medical school, first described the clinical and anatomical features of this congenital anomaly of the tricuspid valve. The patient he studied was a 19 year old labourer who died after suffering increasing symptoms of palpitation, dyspnoea, and cyanosis.<sup>1</sup>

Pregnancy in women with Ebstein's anomaly has been mentioned briefly in a few reports.<sup>2-4</sup> The numbers are small, however, and there is little information on pregnancy and fetal outcome in this disorder. A considerable proportion of women with the anomaly reach childbearing age and it is important that they receive appropriate advice in the reproductive years.

## Patients and methods

Information was obtained by review of medical, obstetric, and neonatal records, and where possible patients were interviewed personally. The 12 women we studied had been referred for assessment to the Prince Charles Hospital, which is the cardiothoracic centre

for the state of Queensland. They include all cases that could be found in the hospital records of women with Ebstein's anomaly who survived infancy. One case was excluded because of other major congenital anomalies (coarctation with corrected transposition of the great arteries).

Electrocardiograms, echocardiograms, and radiological data were reviewed. The anatomical severity of the lesion was determined by echocardiographic assessment and measurement of the ratio of atrialised right ventricle to the total right ventricle (aRV/RV) by Shina's method.<sup>5</sup>

## Results

### PATIENTS

In six women the diagnosis was not known at the time of pregnancy. In one patient the diagnosis was made during her second pregnancy. Two women had associated cyanosis and two had Wolff-Parkinson-White syndrome. The severity of tricuspid valve displacement ranged from a ratio of atrialised right ventricle to right ventricle of 16% to 71%. Only one patient had an associated congenital heart lesion which was pulmonary stenosis.

### PREGNANCY OUTCOME

There were 42 pregnancies. Most of these were well tolerated with 36 live births (table 1). There were five miscarriages (four in patient 12), one ectopic pregnancy, and one neonatal death. Four pregnancies were complicated by premature labour and one by antepartum haemorrhage.

Most infants were born by spontaneous vaginal delivery. Three pregnancies in women known to have Ebstein's anomaly were induced because of maternal dyspnoea. There were four forceps deliveries and three caesarean sections. Epidural anaesthesia was used in only two deliveries, one by forceps and the other by caesarean section. General anaesthetics were given for the other two caesarean sections. No complications occurred with either form of anaesthesia.

Antibiotic cover was given in 12 deliveries where the diagnosis of Ebstein's anomaly in the mother was known, and in two because of the presence of a cardiac murmur. There were no cases of endocarditis.

Most women reported symptoms of dyspnoea in the third trimester of pregnancy. Only patients 3 and 4 were treated with digoxin and none required diuretics. Several women experienced palpitation but there were no

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Table 1 Obstetric and infant outcome in women with Ebstein's anomaly

Patient	Patients					Obstetric outcome					Infant outcome		
	Age	Age at diagnosis (yr)	aRV:RV (%)*	Cyanosis	WPW	No of pregnancies	Live births	SVD	Forceps	Caesarean section	Premature	SFGA	Comments
1	31	7	16		+	2	2	—	1	1	1	—	
2	38	10	53			2	2	1	1	—	—	1	
3	18	16	71	+		1	1	—	—	1	—	—	
4	26	21	50			4	4	2	2	—	2	—	
5	38	24	66		+	4	4	4	—	—	—	—	
6	51	27	29			6	4	4	—	—	—	1	1 miscarriage 1 ectopic pregnancy
7	Dead	48	44			2	2	2	—	—	?	?	
8	57	50	43			4	4	4	—	—	—	1	Pulmonary valvotomy after first pregnancy
9	54	53	47			4	4	4	—	—	—	—	
10	69	56	66	+		2	2	2	—	—	1	1	
11	69	68	42			1	1	—	—	1	—	—	
12	Dead	73	65			10	6	6	—	—	?	?	4 miscarriages, 1 neonatal death

\* Ratio of atrialised right ventricle to right ventricle (aRV/RV) expressed as a percentage (%).

SVD, spontaneous vaginal delivery; SFGA, small for gestational age (no infants were both premature and small for gestational age).

? Information on prematurity and dysmaturity not available.

documented arrhythmias and only patient 1 required treatment.

Two women had considerable problems during pregnancy related to their cardiac anomaly. Case 1 who had Wolff-Parkinson-White syndrome but relatively mild valve displacement was unwell with arrhythmias during both her pregnancies. This was in spite of prior His ablation surgery and anti-arrhythmic treatment. She required a forceps delivery in the first pregnancy because of symptomatic paroxysmal atrial tachycardia in the second stage of labour. In her second pregnancy she required two admissions for control of arrhythmia and cardioversion for resistant atrial fibrillation at 34 weeks followed by urgent caesarean section.

In contrast, the dominant features in case 3 were cyanosis and severe displacement of the tricuspid valve, with a ratio of atrialised right ventricle to right ventricle of 71%. This patient required admission at 26 weeks' gestation with grade III New York Heart Association symptoms of dyspnoea and progressive cyanosis, and she underwent a caesarean section at 36 weeks' gestation with resolution of her symptoms.

#### INFANT OUTCOME

Five of the 36 live born infants were premature (less than 37 weeks' gestation), with birth weights ranging from 1360 g to 2500 g (table 2). Two of the three infants born to mothers with cyanosis were in this group. A further four term infants were small for gestational age (less

than the tenth percentile) including the third infant born to a mother with cyanosis (table 3).

Only one neonatal death occurred (cause unknown) in a twin born at term. All other infants survived and are alive and well and none has congenital cardiac or other anomalies.

#### Discussion

In pregnancy several important physiological changes occur within the cardiovascular system. Plasma volume increases by 40%, reaching a maximum at 30 weeks. A smaller rise in red cell mass of 20 to 30% results in the dilutional anaemia of pregnancy. Cardiac output rises by 40%, with an increase in both stroke volume and heart rate<sup>6</sup> and with an accompanying rise in circulating catecholamine concentrations.<sup>7</sup> However, despite the increase in cardiac output, systemic blood pressure is lower than in the non-pregnant state, indicating a considerable reduction in peripheral vascular resistance due to peripheral vasodilation and placental shunting.<sup>7</sup>

In patients with Ebstein's anomaly, these physiological changes may have appreciable adverse haemodynamic consequences. In the presence of impaired right ventricular size and function, the increased stroke volume may be poorly tolerated and result in worsening tricuspid incompetence, raised right atrial pressures, and increased right to left shunting. Complications of pregnancy and delivery such as haemorrhage may not be well tolerated by these

Table 2 Features and reason for delivery of infants who were born prematurely (less than 37 weeks' gestation)

Gestation (w)	Birth weight (g)	Features and reason for delivery
28	1360	Spontaneous preterm labour
32	1475	Spontaneous preterm labour, maternal cyanosis
34	2540	Urgent caesarean section, resistant maternal arrhythmias
36	2480	Spontaneous preterm labour
36	2170	Caesarean section for worsening maternal dyspnoea and cyanosis

Table 3 Features and reason for delivery of infants born small for gestational age (birth weight less than the 10th percentile for gestation)

Gestation (w)	Birth weight (g)	Features and reason for delivery
39	1815	Spontaneous vaginal delivery, maternal pulmonary stenosis corrected after 1st pregnancy
39	2310	Maternal dyspnoea, spontaneous vaginal delivery
40	2615	No maternal symptoms, forceps delivery for failure to progress
40	2950	Maternal cyanosis, spontaneous vaginal delivery

mothers where right ventricular dysfunction may impair the patient's capacity to cope with the large volume shifts associated with fluid resuscitation.

In the present series, pregnancy was generally well tolerated. Mild dyspnoea was a common symptom in the third trimester but no more troublesome than has been described in normal pregnancy.<sup>8</sup> Only in case 3 did overt symptoms of right heart failure develop. This mother had cyanosis and very severe displacement of the tricuspid valve. However, four patients who also had considerable displacement of the tricuspid valve tolerated pregnancy well. Therefore no correlation was demonstrated between symptoms and the degree of displacement of the tricuspid valve leaflet as described by Shina *et al.*<sup>5</sup>

Patients with Ebstein's anomaly are liable both to supraventricular and ventricular arrhythmias, and Wolff-Parkinson-White syndrome occurs in up to 20% of patients.<sup>9</sup> The raised concentrations of circulating catecholamines in pregnancy will exacerbate any predisposition to arrhythmias and major arrhythmias may occur, particularly if there is maternal hypoxaemia or stress. One of our patients had severe arrhythmias in two pregnancies and required cardioversion despite antiarrhythmic treatment (digoxin, verapamil, flecainide). She had Wolff-Parkinson-White syndrome and in the non-pregnant state had sufficiently severe arrhythmias to warrant His ablation surgery.

Arrhythmias in patients with Ebstein's anomaly, however, are not an absolute contraindication to pregnancy. Various antiarrhythmic drugs have been used without adverse effects on the fetus—for example, propranolol, disopyramide, procainamide, flecainide, sotalol, and amiodarone.<sup>10–12</sup> Patient 5, who had no severe arrhythmias before pregnancy, shows that Ebstein's anomaly with Wolff-Parkinson-White syndrome may in fact be well tolerated. Therefore, advice regarding the risks of pregnancy depends in part on the capacity to achieve arrhythmia control in the non-pregnant state.

Maternal cyanotic congenital heart disease has been shown to be associated with prematurity and low birth weight and with infant survival rates of 50–55%.<sup>2,3</sup> The combined effects of maternal hypoxaemia and haemodynamic instability make the infant liable to intrauterine fetal growth retardation and even death. The three infants born to cyanosed mothers in our study were premature or dysmature, but all survived and did well.

No baby had congenital heart disease in this series. While the numbers are small, the rate of congenital heart disease seems to be low compared with published data rates of 2–14%.<sup>3,13</sup>

Obstetric complications were infrequent in the present series. Only one woman had mild pre-eclamptic toxemia without a significant change in her symptoms. The miscarriage rate of 12% is similar to standard rates of 10% to 15%.<sup>14</sup> Despite the increased cardiovascular stress and major fluid shifts associated with labour and delivery, these final stages of preg-

nancy were well tolerated. Special intervention and monitoring were rarely used. The 7% caesarean rate and 9% forceps delivery rate compare favourably with standard rates of 5–20% and 5–15% respectively. Though antibiotic prophylaxis was given in only 14 of the 42 pregnancies there were no episodes of endocarditis, which accords with the reported low incidence.<sup>3</sup>

There are few published reports on pregnancy in women with Ebstein's anomaly. The largest series was reported by Radford *et al.*, where successful pregnancy occurred in six women who had a total of 12 children.<sup>4</sup> In two large studies on congenital heart disease in pregnancy (total 357 women) five women had Ebstein's anomaly and three of them had cyanosis.<sup>2,3</sup> They had eight pregnancies and there were two fetal deaths, both in infants born to women with cyanosis. Several case reports emphasise the potential complications in pregnancy in women with Ebstein's anomaly.<sup>15–17</sup> However, in each one the patients were symptom free until the third trimester and mother and infant survived.

Though pregnancy was well tolerated in patients in this study and fetal outcome was good, five women including three in whom there were no strong medical or obstetric indications had been advised not to have further pregnancies because of their cardiac anomaly. This uncertainty about the natural history of the condition in pregnancy is reflected in a report of unnecessary termination of pregnancy because of the diagnosis of Ebstein's anomaly in the mother.<sup>18</sup>

The results of this study indicate that women with Ebstein's anomaly who reach childbearing age can be advised that pregnancy is likely to be well tolerated and fetal outcome good. The presence of arrhythmia or cyanosis in the mother is associated with increased maternal and fetal risk and are indications for closer maternal and fetal observation during pregnancy and delivery.

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